

## APPENDIX

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**Appendix Table S1.** The exact P values for Figure EV1

	<b>Figure EV1C</b>	<b>Figure EV1D</b>	<b>Figure EV1E</b>	<b>Figure EV1F</b>	<b>Figure EV1G</b>
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001
wild-type versus treated Lmna <sup>G609G/+</sup>	0.0154	0.0029	0.012	0.0278	0.0001
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	0.0199	0.0352	0.0096	0.0211	0.0001
One-way ANOVA and Tukey's post hoc test					

**Appendix Table S2.** The exact P values for Figure 1

	<b>Figure 1A</b>	<b>Figure 1B</b>	<b>Figure 1C</b>
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0,0001	<0,0001	<0,0001
wild-type versus treated Lmna <sup>G609G/+</sup>	0,0105	0,006	0,0296
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	0,0041	0,0102	0,0074
One-way ANOVA and Tukey's post hoc test			

**Appendix Table S3.** The exact P values for Figure EV2

	<b>Figure EV2A</b>	<b>Figure EV2B</b>	<b>Figure EV2C</b>	<b>Figure EV2E</b>
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	0.0001	<0.0001	<0.0001
wild-type versus treated Lmna <sup>G609G/+</sup>	0.0052	0.1493	0.002	0.0607
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	<0.0001	0.0259	0.0004	0.001
One-way ANOVA and Tukey's post hoc test				

	<b>Figure EV2D</b>			<b>Figure EV2F</b>	
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	<0.0001	<0.0001	<0.0001	0.7051
wild-type versus treated Lmna <sup>G609G/+</sup>	<0.0001	<0.0001	0.0389	0.0215	0.7879
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	0.5409	<0.0001	0.0376	0.0372	0.9892
One-way ANOVA and Tukey's post hoc test					

**Appendix Table S4.** The exact P values for Figure 2

	<b>Figure 2A</b>	<b>Figure 2B</b>	<b>Figure 2C</b>	<b>Figure 2D</b>	<b>Figure 2E</b>
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	<0.0001	<0.0001	<0.0001	0.0023
wild-type versus treated Lmna <sup>G609G/+</sup>	<0.0001	0.0006	<0.0001	0.1232	0.502
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	0.0292	0.0273	0.011	0.017	0.0407
One-way ANOVA and Tukey's post hoc test					

**Appendix Table S5.** The exact P values for Figure 3

	<b>Figure 3C</b>	<b>Figure 3D</b>	<b>Figure 3E</b>	<b>Figure 3F</b>
1 mmol/L Pi versus 2 mmol/L Pi	<0.0001	<0.0001	<0.0001	<0.0001
1 mmol/L Pi versus 2 mmol/L Pi + Mg	0.0014	<0.0001	<0.0001	<0.0001
1 mmol/L Pi versus 2 mmol/L Pi + PPi	0.8959	0.987	0.7854	0.9455
1 mmol/L Pi versus 2 mmol/L Pi + PFA	0.8483	0.9662	0.9811	0.9727
2 mmol/L Pi versus 2 mmol/L Pi + Mg	<0.0001	0.9284	<0.0001	0.9984
2 mmol/L Pi versus 2 mmol/L Pi + PPi	<0.0001	<0.0001	<0.0001	<0.0001
2 mmol/L Pi versus 2 mmol/L Pi + PFA	<0.0001	<0.0001	<0.0001	<0.0001
2 mmol/L Pi + Mg versus 2 mmol/L Pi +PPi	0.021	<0.0001	<0.0001	<0.0001
2 mmol/L Pi + Mg versus 2 mmol/L Pi +PFA	0.0285	<0.0001	<0.0001	<0.0001
2 mmol/L Pi + PPi versus 2 mmol/L Pi +PFA	>0.9999	0.9999	0.9767	>0.9999
One-way ANOVA and Tukey's post hoc test				

**Appendix Table S6.** The exact P values for Figure 4

	<b>Figure 4A</b>	<b>Figure 4B</b>	<b>Figure 4D</b>	<b>Figure 4C</b>
wild-type versus untreated Lmna <sup>G609G/+</sup>	----	----	----	<0.0001
wild-type versus treated Lmna <sup>G609G/+</sup>	----	----	----	0.0196
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	<0.0001	0,0018	0.0034	<0.0001
Unpaired t test			One-way ANOVA and Tukey's post hoc test	

**Appendix Table S7.** The exact P values for Figure EV3

	<b>Figure EV3A</b>	<b>Figure EV3B</b>	<b>Figure EV3C</b>	<b>Figure EV3D</b>	<b>Figure EV3E</b>
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001
wild-type versus treated Lmna <sup>G609G/+</sup>	0.0002	<0.0001	<0.0001	<0.0001	<0.0001
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	0.0334	0.3205	<0.0001	<0.0001	0.2132
One-way ANOVA and Tukey's post hoc test					

**Appendix Table S8.** The exact P values for Figure 5

	<b>Figure 5A</b>	<b>Figure 5B</b>	<b>Figure 5C</b>			
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	<0.0001	<0.0001			
wild-type versus treated Lmna <sup>G609G/+</sup>	0.0034	0.1039	0.3034			
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	0.001	<0.0001	0.0064			
One-way ANOVA and Tukey's post hoc test						
	<b>Figure 5D</b>	<b>Complex I</b>	<b>Complex II</b>	<b>Complex III</b>	<b>Complex IV</b>	<b>Complex V</b>
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	0.374	0.0002	<0.0001	<0.0001	
wild-type versus treated Lmna <sup>G609G/+</sup>	0.0049	0.4839	0.1103	<0.0001	0.0011	
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	<0.0001	0.0108	0.0402	<0.0001	<0.0001	
One-way ANOVA and Tukey's post hoc test						

**Appendix Table S9.** The exact P values for Figure EV4

	<b>Figure EV4A</b>		<b>Figure EV4B</b>	
	0.1 mmol/L	1 mmol/L		
wild-type versus untreated Lmna <sup>G609G/+</sup>	<0.0001	<0.0001	<0.0001	<0.0001
wild-type versus treated Lmna <sup>G609G/+</sup>	0.0024	<0.0001	0.1326	0.0005
untreated Lmna <sup>G609G/+</sup> versus treated Lmna <sup>G609G/+</sup>	0.0068	0.0001	<0.0001	<0.0001
	One-way ANOVA and Tukey's post hoc test			